

Longitudinal Study of the Cognitive Development in Children With Williams-Beuren Syndrome

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Crisco [1990: Clin Res 38:536A] reported stability of IQs in a group of 14 children with Williams-Beuren syndrome (WBS) over a 5-year period and concluded that they display a development rate similar to normal children. The aim of our study was to examine the stability of the development of non-verbal reasoning abilities over a period of 2 years using two methods. We studied 18 children with WBS (9 girls, 9 boys) with a mean age of 6.6 years (range: 4.4–10.6 yr) at year one (T1), and approximately two years later (T2) at the average age of 8.6 years (range: 5.11–12.7 yr). The Columbia Mental Maturity Scale (CMM) and the Draw A Person Test were administered. The results show that the IQs resulting from the Draw A Person Test were stable over the 2-year period (T1: mean IQ = 63.5, T2: mean IQ = 65, $t = 0.63$), and display a significant correlation between the two methods ($r = 0.547$, $P = 0.01$). Furthermore, the correlation between the two tests (CMM and Draw A Person Test) at the second assessment is high and significant ($r = 0.56$, $P = 0.01$). The mean IQs at T2 can be classified as mild mental retardation. A notable result is the significant decrease of the IQs according to the CMM (T1: mean IQ = 77, T2: mean IQ = 68, $t = 2.69$, $P = 0.01$). These results suggest that the developmental outcome of children with WBS varies in specific areas of cognitive function over a 2-year period.

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INTRODUCTION

The most important manifestations in the Williams-Beuren syndrome (WBS) are the distinct facial appearance, cardiovascular defects, renal anomalies, growth delay, and mental retardation [Beuren et al., 1962; Morris et al., 1988; Pankau et al., 1992; Pober et al., 1993; Williams et al., 1961]. Depending on the kind of study and the sample size, the mental retardation ranges from an average IQ between 40 and 79 [Sherrets et al., 1982; Gosch and Pankau, 1994]. The retardation can be classified as mild to moderate [cf. DSM-IV, American Psychiatric Association, 1994].

In his study, Crisco [1990] examined the stability of global IQ in 14 children with WBS over a period of 5 years. At the first examination the average age of the children was 50.36 months and the mean intelligence score assessed by the Stanford-Binet Intelligence Scale, Form L-M, was 67 (S.D. = 16.53). Five years later, the children were 111.07 months old on average and their mean IQ was 66 (S.D. = 11.88). The difference was not statistically significant. Crisco concluded that children with WBS show lower ability levels but “appear not less stable in developmental rate than normal children.”

There have been no further studies that confirm the findings of Crisco [1990]. The aim of our study was to examine different aspects of cognitive development in children with WBS over a period of 2 years.

METHODS

We investigated 18 children with WBS (9 girls and 9 boys). The children were tested twice over a period of approximately two years. The mean ages were 6.6 years (range: 4.4 to 10.6 years) and 8.6 years (range: 5.11 to 12.7 years). They were examined either at the Department of Pediatrics, University of Kiel, or during the 2nd or 3rd Meeting of the Federal German WBS Association.

Two measures were administered to the children to study their nonverbal reasoning abilities: first, the Columbia Mental Maturity Scale-CMM [Bondy et al., 1969]. This test has shown a high reliability (internal consistency according to Kuder-Richardson $r = 0.97$ for retarded children), test-retest reliability (split-half: $r = 0.83$ – 0.96 for normally developed and $r = 0.97$ for mentally retarded children), external validity (for instance, intercorrelations to the German WISC: $r = 0.49$ for mentally retarded individuals), and discriminates be-

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tween normally developed and mentally retarded children. Secondly, the Draw A Person Test, which is a psychometric measure to assess the children's intellectual maturity and their concepts of the human figure as an index of their general concepts, was administered. The basic research on this test was done by Florence Goodenough [1926], but in our investigation the children's drawings were evaluated according to Ziler [1971].

Statistical analysis was performed using t-tests for dependent random samples, and Pearson-Product-Moment-Correlations were computed between the two points of time to determine the degree of individual stability for the two tests. The probability that observed differences were due to chance was determined by Bonferoni correction of the significance level, P values < 0.05 . The computer version of the "Statistical Package for Social Sciences SPSS" was used to calculate the statistical procedure.

RESULTS

Subjects

All subjects lived at home, and 94% ($n = 17$) were brought up by both parents, while one child was raised by the mother alone. Only 11% of the children with WBS had no sibs; most of the others (67%) had one more sister or brother and there were more than two sibs in the remaining families. The socioeconomic level was rated by the educational level and occupation of the father [cf. Bolte et al., 1975]; 17% ($n = 3$) of the families belonged to the upper class, 55% to the middle and 22% to the lower class.

Six of the children with WBS attended normal kindergarten, 10 an integrative group or a special kindergarten and on 2 children we had no information. Additionally, most of the children experienced about 3 therapies (cf. Fig. 1).

Psychological Results

Table I shows that when first (T1) assessed by the Columbia Mental Maturity Scale (CMM) the children with WBS had an average IQ of 77 (S.D. = 14.7) while two years (T2) later the mean IQ was 68 (S.D. = 13.1). In comparison, the mean IQ by the Draw A Person Test at T1 was 63 (S.D. = 13.2) and at T2 65 (S.D. = 10.7). T-Test for random dependent samples between time T1 and T2 was not significant for the Draw A Person Test ($t = .63$, $P = .54$), but for the CMM ($t = 2.7$, $P = .01$). This means that children with WBS displayed a decrease in performance over a 2-year period according to the CMM.

The Pearson-Product-Moment-Correlation computed over time for both measures indicated significant correlations for both the Draw A Person Test ($r = 0.55$, $P = 0.01$) and CMM ($r = 0.55$, $P = 0.01$). Additionally, the correlation between the IQs of both tests at the first evaluation T1 is lower and not significant ($r = 0.43$), while the correlation between the two tests at T2 is higher and significant ($r = 0.56$, $P < 0.01$).

The changes in the IQ in the children are presented in Figures 2 and 3. We categorised the children into three groups: normal IQ: ≥ 85 ; learning disability IQ: 84-70; mental retardation IQ: ≤ 69 .

Figure 2 shows the changes of IQ as assessed by the CMM. The percentage of the group of children with a learning disability did not change. In comparison, fewer individuals could be classified as having a normal IQ, and a higher percentage of children showed mental retardation after two years. This means that an increase from 22% to 50% of the children belonging to the group of mental retardation at T2 can be seen.

In Figure 3 the changes according to the results of the Draw A Person Test are presented. A decrease in the group of mental retardation and an increase in the group

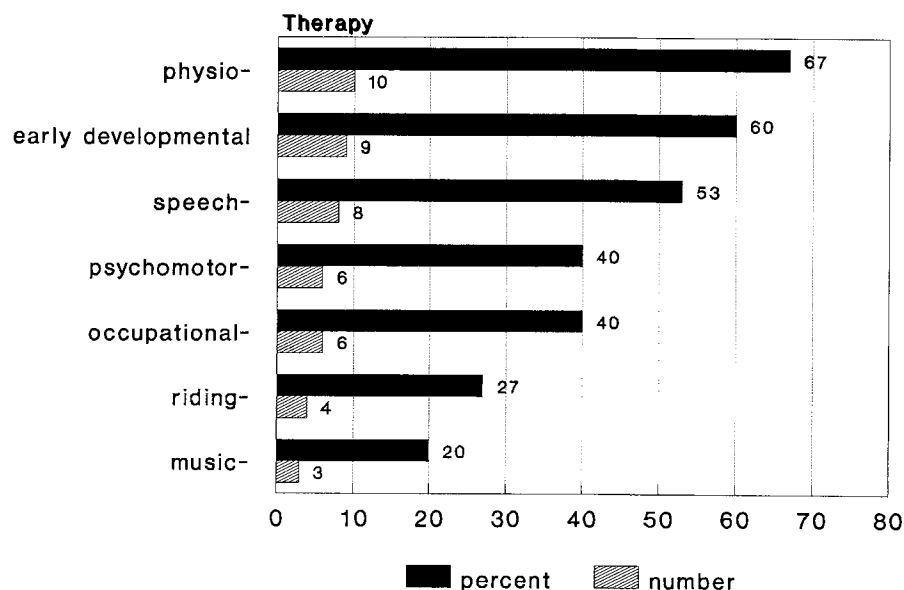


Fig. 1. Number and percentages of the 18 children with WBS receiving special therapies.

TABLE I. Changes of Mean Scores and Standard Deviations of the IQs in Children With WBS Investigated by the Columbia Mental Maturity Scale (CMM) and Draw a Person Test (DAP)

	T1 ^a		T2		t	P ^d
	M ^b	S.D. ^c	M	S.D.		
CMM	77	(14.7)	68	(13.1)	2.69	.015
DAP	64	(13.2)	65	(10.7)	-.63	.537

^aT1: first assessment; T2: second assessment approximately two years later

^bM, mean.

^cS.D., standard deviation.

^dP, level of significance.

of learning disability was observed, while the percentages in the group of normal IQ did not change.

DISCUSSION

Our results show that IQs of children with WBS assessed by the Draw A Person Test are stable over a 2-year period, whereas data resulting from the CMM indicate a decrease of performance.

The Draw A Person Test is a very economical and quick method. Although children with WBS show significant difficulties in their fine motor coordination, this seems to be an adequate test to estimate their cognitive nonverbal abilities and concepts of the human figure. The IQs were stable over time, and when tested two years later (T2) there was a significant correlation between the IQs obtained from the Draw A Person Test and the CMM. Additionally, these results are in accordance to those reported by Crisco [1990].

The decrease in IQ measured by the CMM over the 2-year period is surprising. This raises the question whether the norms for the German version of the CMM

are not adequate for younger children, or overestimate their nonverbal reasoning abilities, or whether this test investigates aspects of intelligence in which a decrease, or a slower developmental rate than expected, occur. To answer this question we examined the children's results from this test in detail. In the CMM the children have to differentiate and decide which characteristic from a number of signs or figures is different. The test consists of concrete or abstract objects, and either there is one false object in a series (for example item no. 24: three houses and one horse) or classifications have to be made by the child (for example item no. 40: two elephants, two chairs and one cup) to solve the task. It can be hypothesized that children with WBS display more difficulties in solving items with abstract material and in forming classifications than with problems involving concrete objects. Therefore, we calculated the percentage of correctly answered items with reference to the total number of concrete or abstract items. The descriptive evaluation of the single T2 answer sheets showed that children with WBS solved more concrete items (median 79%) correctly than abstract material (median 68%). They also solved more items correctly when asked to find one false object out of a series of features (median 53%) in comparison to classification (mean 33%). This shows that it is easier for children with WBS to produce right solutions when the material is concrete. It may be easier for the children because they are able to use verbal strategies to find the right solution. Further, the WBS individuals solve fewer items correctly when they have to make classifications, which means having to differentiate between two characteristics to find the right answer. In comparison to T1, the children are expected to be able to solve many of these more difficult items at T2. We suppose that the decrease in the IQ at T2 was due to this fact.

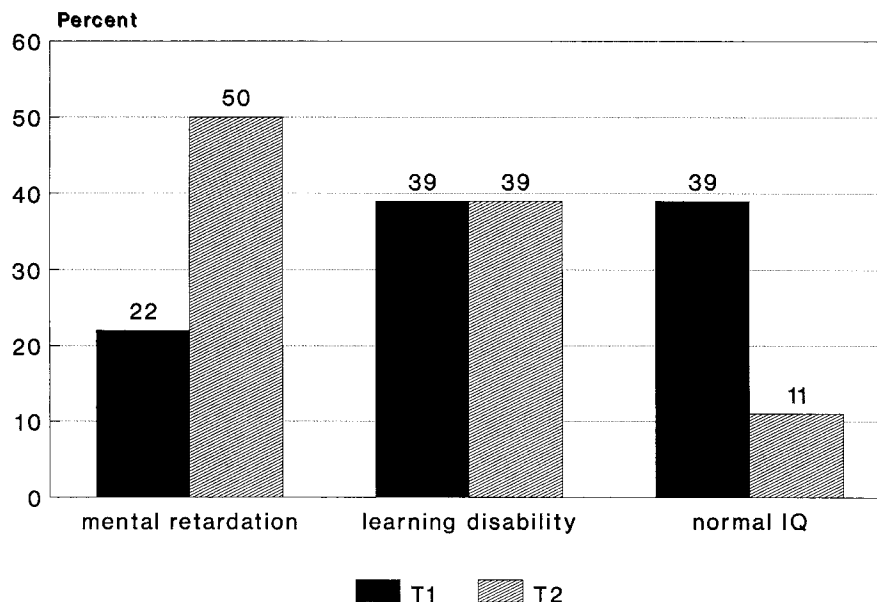


Fig. 2. Percentages of children with WBS according to IQ groups measured by the CMM over a 2-year period.

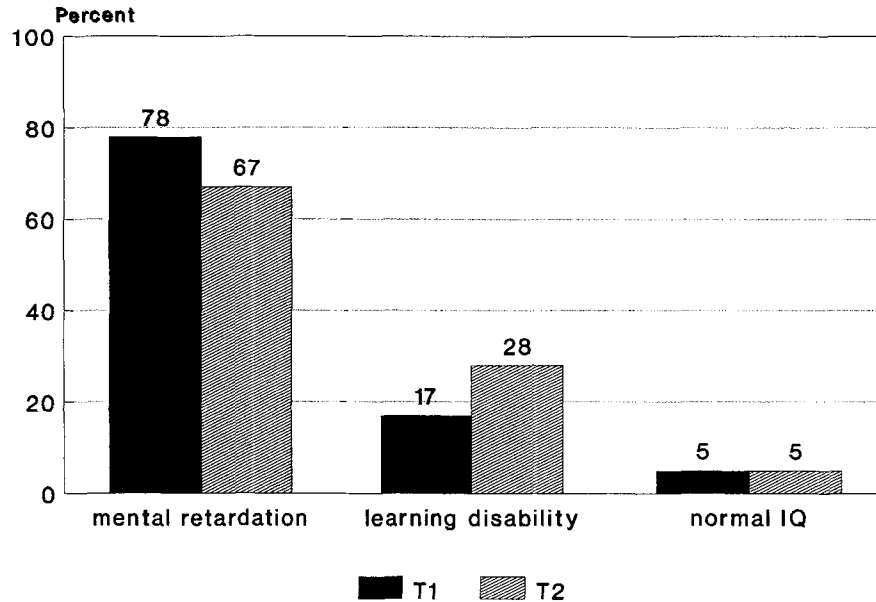


Fig. 3. Percentages of children with WBS according to IQ groups measured by Draw a Person Test over a 2-year period.

The children with WBS experienced an average of three therapeutic programs (speech, psychomotor therapy, etc.). Unfortunately, the frequency, duration, efficacy of the treatments are unknown in this group. Different assumptions about the influence of a therapy on the developmental outcome in specific areas of cognitive function are possible. It can be speculated that a therapeutic program influences the cognitive outcome directly (positively or negatively) or indirectly (e.g., increases the attention span of children with WBS or helps them to concentrate longer, with the result that they have a better opportunity to learn more). It would be helpful to investigate the effects of different therapeutic intervention programs on the cognitive performance of children with WBS. One problem with this research approach is that a control group without any intervention would be needed, which is not possible for ethical reasons.

Finally, our results are in accordance with those of Crisco [1990]. That means a high stability of nonverbal intelligence quotients in children with WBS at the level of mild mental retardation can be found. Additionally, in our study we were able to show that, depending on the test employed, a high stability of the IQ over a 2-year period could be observed, as well as a decrease in the development rate of special abilities such as the application or use of classifications. In future studies the development of different domains of cognitive ability should be investigated more thoroughly.

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